



Challenging Behaviors in Huntington's Disease: Strategies for Patients and Families

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Huntington's disease (HD) is a *neuropsychiatric* illness, which means that people with HD suffer from both physical symptoms, like chorea, and mental symptoms, such as depression or problems with thinking and memory. Research tells us that psychiatric symptoms in HD often arise years earlier than physical symptoms, and that psychiatric symptoms are frequently the most distressing symptoms for both patients and families to cope with. This article provides an overview of behavioral symptoms common in HD, and suggests strategies for how to best manage them.

Psychiatric problems are extremely common in those suffering from Huntington's disease; in fact, almost all patients with Huntington's disease will have at least one psychiatric symptom during their lives, and most patients will have multiple symptoms. Behavioral symptoms in HD are a direct result of changes in the brain caused by the illness. This happens because Huntington's disease damages important structures and pathways in the brain—and this damage causes the problems with movement, thinking, and behavior we see in Huntington's patients. Though the psychiatric problems common in HD can be challenging for patients and families, it's important to remember that these problems are *caused by the illness*, and not by the person suffering from Huntington's disease. Additionally, it's crucial to keep in mind that people with HD face many losses, including loss of independence, health, and eventually, their lives. These losses burden HD patients with frustration, anger, and grief in addition to any psychiatric symptoms they may have from the illness. Understanding that behavioral issues in HD stem from a combination of brain changes and the effects of profound loss helps to guide coping strategies for patients and families. The following table summarizes common behavioral symptoms in HD.

Common Challenging Behaviors in Huntington's Disease

Unawareness: Failure to recognize or notice problematic behaviors or the declining ability to function.

Examples: Person doesn't notice worsening performance at work, person fails to recognize they are no longer a safe driver.

Declining executive function: Problems with speed of thinking, planning, prioritizing, organizing, concentration, decision-making, flexibility, or creativity.

Examples: Poor performance at work, inappropriate behavior, impulsive decision-making

Apathy: Loss of motivation to attend to responsibilities, basic needs, and things the person used to think were important.

Examples: Difficulty getting out of bed or starting the day, neglecting household chores or personal hygiene

Irritability and disproportionate anger: Often stems from frustration about losses (e.g. abilities, independence) combined with brain changes that decrease the ability to regulate emotions.

Examples: screaming, swearing, threatening, slamming doors, hitting walls, pushing, striking or hurting others

Obsessive thoughts and compulsive behaviors: Recurrent, intrusive thoughts paired with repetitive behaviors that reduce inner discomfort.

Examples: Over-concern with germs/contamination, fixation on past insults/injustices (thoughts); repeated hand washing, compulsive eating or drinking (behaviors)

Depression and suicide: pattern of low mood and poor energy that can lead to feelings of hopelessness and thoughts about ending life.

Signs: Persistent sadness, irritability, or low energy; changes to appetite and sleep patterns; preoccupation with or frequent thoughts about death or suicide

Anxiety: Intense feelings of inner discomfort, worry, panic, or restlessness.

Signs: Frequent worrying about minor or everyday things, fear of losing control or "going crazy," preoccupation with perceived judgment or scrutiny from others, obsessions/compulsions (see above).

Strategies for problems with executing functioning and apathy

Problems with executive functioning (thinking, planning, and organizing) and apathy (loss of motivation to initiate or complete activities) can significantly impact the lives of people with HD and their families. Fortunately, similar strategies are helpful for managing both of these problems. HD patients with apathy or deficits in executive function may benefit dramatically from *having a regular routine*. Having a predictable schedule (e.g. regular meal and bed times, chores completed at same time each day, etc.) is helpful because it reduces the amount of new information and change the person with HD faces. This is especially helpful for individuals experiencing problems with thinking and memory. The familiarity of a routine is also soothing for some, and can reduce anxiety and irritability. Additionally, patients and families may find the use of *cues and prompts* helpful. Cues and prompts help the person to remember and pay attention to his or her responsibilities and routine. Examples include using cell phone alarms, calendars, and gentle verbal prompts to remind individuals with HD that it's meal or medication time, or that a chore needs completing. *Additional helpful strategies* include speaking in short sentences that give 1-2 pieces of information at a time, or offering choices instead of open-ended questions (e.g. "Do you want oatmeal or eggs?" instead of "What do you want for breakfast?").

Strategies for inappropriate anger and unawareness

Sometimes, individuals with HD may experience inappropriate anger that seems out of proportion to the situation at hand. Individuals may lose patience more quickly than they used to, or may not be able to “shrug off” minor irritations. The person’s anger may escalate quickly, and they may yell, slam doors, throw objects, or even hurt others. This can be especially challenging if the person with HD also suffers from unawareness, or the inability to recognize their symptoms or declining functioning. One important strategy is to *avoid direct confrontation*, and adopt a helpful stance when possible. For example, instead of saying “You can’t drive because you have Huntington’s—your attention and motor skills aren’t good,” try, “I’ll drive you—I was going there today anyway.” This helps the person to maintain a sense of dignity and control, and is less likely to trigger anger. Another critical strategy for preventing anger outbursts is for the person with HD to *stop all use of alcohol and recreational drugs*. Though someone using these substances may feel better in the moment, substance use increases the risk of dangerous behavior and is not safe for people with HD. For particularly sensitive issues, such as approaching the person about declining work performance or unsafe driving, *involving outside agencies* is helpful (e.g. work performance evaluation, driver evaluation through DMV) because it shifts confrontation away from family members. If the person suffering from HD does become inappropriately angry, *de-escalation techniques* such as using soft voice, kind words, and giving space may help. If the person threatens or uses violence, it is crucial to *get away and call for help* (e.g. police). Do not attempt to touch or restrain the person yourself. Any HD patient with frequent or severe inappropriate anger should see their HD care team right away, because they may need *medications* to help reduce anger. *Additional important strategies* include minimizing stress, removing any weapons from the home, and maintaining a calm, predictable environment whenever possible.

Strategies for depression, anxiety, and other psychiatric syndromes

In addition to the above behavioral challenges, people with HD often suffer from distinct psychiatric syndromes, such as depression and anxiety. The presence of these types of syndromes can worsen challenging behaviors, so recognizing and treating them is critical. Treatments may consist of medications, counseling or psychotherapy, and other interventions. In addition, some of the above strategies, such as maintaining a consistent routine, can be helpful for depression and anxiety. Lastly, it is important to be aware that *suicide is unfortunately common* among those who suffer from HD. Suicidal thoughts and statements in patients with HD should always be taken seriously and addressed immediately. *If someone with HD voices an intent or plan to commit suicide, it is an emergency*, and those around them should take actions such as calling a suicide hotline, or 911.